NEW YORK NEUROLOGICAL SOCIETY.

March 5, 1901.

The President, Dr. Joseph Collins, in the chair.

A CASE FOR DIAGNOSIS.

Dr. Joseph Collins presented a boy of ten years who had a complex of symptoms which could not easily be placed under any one designation. He was one of twelve children, seven of whom had had in infancy marasmus or gastro-intestinal disorders. The present ailment had begun about nine months ago, at which time the boy had commenced to "hop." He complained of pain in the great toe of the left side, and also of pain in the precordial region. He had then been taken to the Mt. Sinai Hospital, and while there some stiffness or weakness in the lower limbs on walking had been observed. This impairment of motion had steadily increased, so that at the present time he was practically unable to walk more than a few steps. According to the history great difficulty in commencing the act of micturition had been experienced. At present there were no symptoms referable to the bowel or bladder. He has a peculiar waddling gait, and when standing, there is a typical flat-foot. There is a peculiar knocking together of the thighs. The spasticity of the gait had been found, on closer examination, to be more apparent than real. There is a marked ankle-clonus, but no sensory disturbances. The boy gets up from the reclining postion as children do in the early stages of progressive muscular dystrophy. These symptoms, the speaker said, seemed to point distinctly to a lesion in the spinal cord in the crossed pyramidal tracts.

Dr. Sachs said that he had been much interested in this boy at the time he had been in the hospital. The combination of the waddling gait, so characteristic of muscular dystrophy, with an increase in the reflexes, seemed to be very unusual. When the boy was stripped, it seemed to Dr. Sachs very evident that he had progressive muscular atrophy. In addition to this he thought there was a subacute myelitis, possibly of traumatic origin. There could be no question that the calves were hypertrophied. This diagnosis had been arrived at only after careful observation for a period of several weeks. The frequent falls which such children have, would easily explain the occurrence of a subacute myelitis. The spasticity had been more marked nine weeks ago.

Dr. Graeme M. Hammond said that from the cursory examination that he had made he felt inclined to accept the diagnosis given by Dr. Sachs.

Dr. William M. Leszynsky said that it seemed to him to be a very atypical case. He had seen quite a number of patients in whom there had been complete spasticity, and yet the ankle-clonus had been very marked.

Dr. Josèph Fraenkel said that the most prominent symptoms were those of disease of the lateral and posterior columns. At first sight the gait was that of ataxic paraplegia. On the supposition that the myelitis was the later disease, one would expect less evidence of the second. The evidences of pseudo-hypertrophy were certainly quite trifling, while the disease of the cord was very marked. He inclined to the diagnosis of a subacute multiple sclerosis of a paraplegic type.

Dr. J. Arthur Booth said that there was certainly a greater deposit of soft tissue in the left than in the right calf. He accepted the diagnosis given by Dr. Sachs.

Dr. Collins thought it was safe to assume that the boy had a dystrophy, but not that there was a subacute myelitis. No one could say whether the changes in the spinal cord which accompany the dystrophies are not of such nature that double ankle-clonus and double knee-jerk might not develop. It was certainly a form of dystrophy which did not conform strictly to the description of any usually given. If there were an implication of the cord it was confined entirely to the crossed pyramidal tracts. He would not be willing to admit that there were any changes in the spinal cord except secondary ones.

PROGRESSIVE MUSCULAR DYSTROPHIES WITH A REPORT OF A POST-MORTEM EXAMINATION.

Drs. B. Sachs and Harlow Brooks presented this paper. The authors stated that it could not be denied that there was any sufficient distinction between the amyotrophies and the dystrophies. In former years much stress had been laid on the muscular structure. Hypertrophied fibers were found in abundance in dystrophies, whereas in the amyotrophies these fibers were not found. But later it had been shown that the hypertrophied fibers were found in other diseases than dystrophies. It was also a question whether the gray matter of the cord was affected in the primary dystrophies. The case to be reported was one of progressive muscular dystrophy of fifteen years' duration, yet the structural changes, as demonstrated by the latest methods of staining, were very slight. The patient had been admitted to the Montefiore Home eleven years ago at the age of twelve years. Early in life the parents had noticed peculiar movements of the head and eyes. He had been in good health up to about the age of ten years, when he had fallen and broken his leg. At the age of twelve years, after an attack of typhoid fever, the calves were observed to be decidedly hypertrophied. The head was enlarged and exhibited certain movements. There was a marked atrophy of all the muscles of the shoulder-girdle, arm and forearm. The deep spinal muscles were intensely atrophied. The thigh muscles were atrophied. The case became an extreme illustration of a progressive muscular disease of the pseudo-hypertrophic type. The lad's intelligence was fair.

Dr. Brooks said that at the autopsy the organs were normal with the exception of an acute pneumonia and slight myocarditis. There were no gross lesions of the brain or spinal cord. No lesion of the smooth voluntary muscular tissue could be found anywhere in the body. The psoas muscle showed extensive fibrosis. All the muscles of the back showed extensive fibroid replacement, and in places there was a replacement by yellow fat. The trapezii were very extensively invaded. The most extreme changes were in the muscles of the calves, where normal muscular tissue was lost. The autopsy had been done twenty-four hours after death, and at that time there had been no evidence of post-mortem decomposition. On microscopical examination, the muscles showed extensive replacement with areolar tissue of the adult type. In the calf, occasional remnants of voluntary muscle were found. Most of the fibers of the psoas muscles were either larger or smaller than normal. The course striæ could usually be made out. The changes in the other voluntary muscles were of the same character, though varying in extent. In the occipital muscles the amount of connective tissue hyperplasia was less, but nuclear proliferation was prominent. Examination of various portions of the smooth muscles failed to show degeneration or hyperplasia of the connective tissue forming its framework. The heart muscle showed much less connective tissue increase than had been expected from the gross examination. The cardiac muscle was in a very natural condition, there being no atrophy, no abnormal pigmentation or abnormal nuclear activity. The blood vessels in the various tissues showed uniformly an increase in the connective tissue. No evidences of new vessel formation were found. Numerous peripheral nerves were examined, but no appreciable degeneration of fibers was discovered. Only a few of the spinal ganglia had been properly prepared for examination, but these few showed a shrinkage of the ganglion cells similar to that produced by fixing agents. The irregular perilymphatic spaces were, however, found filled with proliferating capsular cells, apparently indicating that this was not an artefact, but a distinct pathological process. Apparently the connective tissue of the ganglia had been increased. The connective tissue throughout the entire cord was found to be increased. The blood vessels of the cord were universally congested, but this was apparently of a hypostatic nature, due to the position of the patient before death. Nothing in the nature of a systemic degeneration of the fibers was found at any level. In the cervical region the ganglion cells in the anterior horn showed a slight nuclear eccentricity. The dendrites universally retained their power to respond to the stain. Occasionally the achromatic elements stained to a slight degree. Eccentricity of the nucleus was found more commonly in the dorsal cord than elsewhere. Lesions in the cells of the posterior horns were more infrequent than in the anterior horns. The most common lesion was a finely granular subdivision of the plaques, usually not involving the entire cytoplasm. A few of the lumbar cells showed an unusual amount of brown pigment collected about the nucleus.

The chief lesions were: (1) Extensive atrophy, which affected apparently all of the voluntary skeletal muscles and was confined to these muscles; (2) the production of areolar connective tissue and adipose; (3) slight general perivascular hyperplasia; (4) moderate interstitial myocarditis; (5) extensive degenerative changes in a few of the posterior root ganglia, and (6) rare changes in the cytoplasm of the ganglion cells of the spinal cord.

The complete absence of the changes in the smooth muscles showed that the disease process was strictly localised in the voluntary muscular system. The authors did not look upon the connective tissue increase as an essential feature of the pathological process, but as an example of a universal function of this tissue to take the place of any tissue which had been removed. The perivascular connective tissue hypperplasia was very slight, and could not be considered as typical of the disease or as produced by it. Possibly the moderate myocarditis was associated with the connective tissue hyperplasia of the blood vessels. It did not seem to be in any way connected with the factors producing atrophy of the voluntary muscles. The changes in the posterior root ganglia seemed to be of great significance, though it was not clear that they bore any direct relation to the changes in the voluntary muscles. These degenerations seemed to be secondary in their nature, and dependent upon death or disease of certain portions of the neurone. A process similar to this occurred after amputation. There were, therefore, no evidences of tract disease. The cytoplasmic degeneration of the ganglion cells in the cord were rare, and might represent the early stage of post-mortem change. Dr. Sachs said that these findings did not indicate that the cause was to be found in the gray matter of the cord. The disease represents a primary affection of the muscular fiber. The occurrence of stigmata of degeneration in so many cases of this dystrophy would lead one to think that these should be broadly classified under family affections. The question arose as to whether these muscular dystrophies were essentially progressive, and the statement was made that in every case the possibility of great improvement by systematic exercise should always be kept in mind in the early stage. Two illustrative cases were briefly reported which had been followed for many years.

Dr. C. L. Dana said that so far as the dystrophies were concerned, which were not strictly of the so-called pseudo-hypertrophic type but rather of the arm and leg type, it seemed to be a well-known fact that many of them cease to progress and patients live years in comparative comfort. He had personal knowledge of two families in which there were six or seven persons, belonging to three generations, who were afflicted with the leg or arm type of dystrophy. Some had lived to old age with only an inability to use the upper arm or perhaps the thigh muscles. One of these patients, when thirty-nine years of age, had been seen at many clinics in New York. The atrophies had begun at the age of nineteen, and had reached their height at about the age of twenty-nine. His weight had been reduced to eighty-eight pounds. A fairly hopeful prognosis could be given in this class of cases, especially when the atrophies do not begin very early in life. In Dr. Dana's experience with pseudo-muscular hypertrophy, there had been only one case in which the disease had been really checked. This person was a lady of twenty-three, in whom the trouble had begun at the age of eighteen. She had presented all of the typical symptoms of pseudo-hypertrophy of late development. He had put her upon systematic exercises, and as a result the disease had not only ceased to progress, but she had absolutely improved. In another case which had been faithfully treated by exercise and massage for four years, there had been continued progress. Dr. Dana said he would like to have Dr. Brooks explain why there should be so much fibrosis in these cases. It might be that in the death of the muscle fiber an irritant poison is formed, and that this gives rise to the increased proliferation. There was a striking difference clinically between a typical spinal atrophy and an ordinary dystrophy, and he believed these diseases were very different in their origin also. The hereditary cases were of a type which is quite distinct from that of the acquired forms.

Dr. A. Wiener said that about six years ago he had presented to the Society, a patient who was very much crippled in his muscular movements. In that case he had been so convinced that many muscular fibers were still intact that he had carefully trained him in exercises for six months. He had been again presented to the Society at the end of that time, and the improvement had certainly been very marked—indeed, the day previous he had been able to ride forty miles on his bicycle. On the other hand, in the dystrophies occurring early in life, he did not believe much could be gained by exercise.

Dr. Leszynsky exhibited photographs of two patients whom he had had under his care for a number of years, with pseudo-hypertrophy of the lower extremities, and progressive muscular atrophy of the upper extremities. Both had died of some intercurrent disease at the age of twenty-five. Before he had seen them, both had received massage and careful training in physical exercise.

Dr. Hammond said that he had recently made a post-mortem examination upon a case of pseudo-hypertrophy, with rather interesting findings. In the lumbar enlargement of the cord, there had been found a distinct cavity extending for several inches. In the dorsal segment was a large gliomatous mass pressing upon the cord. This was apparently a lesion independent of the disease in question. He was in a position to agree entirely with Dr. Wiener, as to the beneficial effect of exercise. He had tried systematic exercise in those affected early in life, and had not found the slightest benefit from carefully conducted exercise of this kind, carried on for a considerable time.

Dr. B. Onuf said that the muscles which have to do the most work were the ones most affected; the muscles which must bear the weight of the body are the first ones to become affected. It was because of this fact that he had been led to try the effect of the opposite plan of treatment, *i.e.*, rest in bed. However, the result had been disappointing.

Dr. Collins said that the post-mortem examination referred to by Dr. Hammond, had been on a patient that had been under his own observation for over two years. This man had been twenty-four years of age at the time of his death, and the disease had existed for fourteen years. Careful investigation into the family history had shown no similar case. Up to his eleventh year he had been free from all disease. For eleven years he had been unable to walk, and for eight years had been bed-ridden. Up to a short time before death he had retained the ability to move his finger, though he could not move his arm, and his facial muscles continued to act well. It was a most typical case of progressive muscular dystrophy. Death had resulted from a tuberculous infection. The brain reported by Dr. Sachs had been from a boy under his observation for six or seven years. Assiduous attention to calisthenic exercises had made him worse in proportion to the assiduity, and that, too, although he had had an excellent calisthenic teacher. This had been Dr. Collins' experience in all other cases in which the disease had begun early in life. The changes described by Dr. Brooks, as having taken place in the ganglion cells, certainly opposed one's ideas of the relation of the neurone to disease. After five years' experience with the Nissl stain, he felt in a position to assert positively that nuclear eccentricity means absolutely nothing.

Dr. Smith Ely Jelliffe said that he had heard nothing said by Dr. Brooks about the terminal end plate of the motor nerve.

Dr. Brooks replied that after attempting to study end plates, he had come to the conclusion that this could only be done with methylene blue during life, and that he had been unable to do. He could not tell Dr. Dana why the connective tissue should grow up so much more in one place than in another. In the calves there had been an increase in volume, and in the pectoral muscles a decrease. It was not improbable that it was due to some inflammatory or toxemic condition in these particular muscles, or possibly that the muscle had been over-exercised. He agreed with Dr. Collins about the uncertainty of the Nissl stain, and that the findings described by him as having been observed in the ganglion cells were negative.

Dr. Sachs closed the discussion. He admitted that the findings reported in the paper certainly did tend to shock one's confidence in the neurone theory as a whole.